American Journal of Hematology 58:100–104 (1998)

Splenic Abscess and Sickle Cell Disease

Ahmed H. Al-Salem,[1]* Syed Qaisaruddin,[1] Ali Al Jam'a,[2] Jaffar Al-Kalaf,[2] and Ali M. El-Bashier[3]

[1]Division of Pediatric Surgery, Department of Surgery, Qatif Central Hospital, Saudi Arabia
[2]Department of Internal Medicine, Qatif Central Hospital, Saudi Arabia
[3]Department of Microbiology, Qatif Central Hospital, Saudi Arabia

This is a report of our experience with 10 cases of splenic abscess in patients with sickle cell disease (SCD). All presented with fever and abdominal pain and were found to have a tender enlarged spleen. Two were found to have a ruptured spleen and five of them were septicemic on presentation. Although both ultrasound and CT-scan of the abdomen were of diagnostic value, we found CT-scan more accurate and reliable in the diagnosis of splenic abscess. Ultrasound and/or CT-scan should be used routinely in the evaluation of SCD patients who present with fever and abdominal pain, especially if they have a tender enlarged spleen. Diagnostic aspiration under CT-scan or ultrasound guidance should be used in doubtful cases to differentiate between splenic abscess and a large splenic infarct. All our patients were managed by peri operative antibiotics and splenectomy with no mortality. Salmonella was the commonest causative organism. Although CT-guided aspiration of splenic abscess is being advocated recently, we feel splenectomy should be the treatment of choice in patients with SCD as there is no point in preserving a non-functioning spleen that is present in the majority of patients. CT-guided aspiration may be employed as a temporary measure for those patients who are at high surgical risk with unilocular abscess. Am. J. Hematol. 58:100–104, 1998.

Key words: splenic abscess; sickle cell disease; salmonella; splenectomy

INTRODUCTION

The signs and symptoms of splenic abscess are generally non-specific leading to delay in diagnosis and treatment with increased mortality, which has been reported as high as 40% [1,2]. However, with the recent advances and liberal use of diagnostic techniques such as ultrasonography and CT-scan, there is an increase in the early detection of splenic abscess with improved outcome, and emergence of other modalities of treatment for splenic abscess, other than splenectomy [3–8]. Recently, there is also a change in the spectrum of splenic abscess with more cases being described in immunocompromised patients [9–11], as well as emergence of unusual causative organisms [8,12–14].

Splenic abscess has always been reported to be uncommon, accounting for 0.14% to a 0.7% of necroposy specimen [15]. This is attributed to the fact that the spleen, which act as a blood filter for bacteria and other organisms, is very resistant to infection. This function is, however, lost or altered in diseases such as sickle cell disease (SCD). These patients are known to develop splenic infarction and are at increased risk of infection especially in the presence of functional asplenia, which predisposes them to the development of splenic abscess [16–18]. This report describes our experience with 10 cases of splenic abscess in patients with sickle cell disease.

PATIENTS AND METHODS

Over a 7-year period, June 1989 to June 1996, a total of about 2,000 patients with SCD were treated at our hospital. Ten of these had splenic abscess. The charts of these patients were reviewed for age at diagnosis, sex, presentation, laboratory and diagnostic studies, management, pathology, and culture results as well as outcome. The diagnosis of SCD was made based on a positive

*Correspondence to: Dr. Ahmed H. Al-Salem, P.O. Box 18432, Qatif 31911, Saudi Arabia.

Received for publication 10 April 1997; Accepted 14 January 1998

TABLE I. Demographic and Clinical Data

No.	Age (years)	Sex	Temp. (°C)	Signs and symptoms	Splenic size (cm)
1	21	M	38	Admitted with Enterobacter Sakazaki septicemia. Treated for 3 weeks and discharged. Readmitted 2 days later with fever and left hypochondrial pain. Found to have tender enlarged spleen.	18 × 8
2	11	M	38.5	Abdominal pain, fever, vomiting, and pain in left upper limb. Developed salmonella septicemia. Found to have enlarged tender spleen.	12.4 × 6.7
3	15	M	38	Admitted with back pain, pain in the legs, fever, and abdominal pain. Developed salmonella septicemia and acute chest syndrome. Found to have enlarged tender spleen.	18 × 7.7
4	26	F	37.8	Admitted with generalized pain all over the body and weakness with fever. Developed salmonella septicemia. Found to have enlarged tender spleen.	14.5 × 6
5	17	M	38.5	Admitted with fever, left back pain, and fullness. Found to have a tender left renal area and tender enlarged spleen.	15 × 8
6	4	M	38.6	Admitted with fever and abdominal pain. Found to have enlarged tender spleen and left basal crepitations.	16.5 × 7.5
7	18	M	38.3	Admitted with upper left quadrant abdominal pain and fever. Found to have tender enlarged spleen.	15.5 × 7.6
8	14	F	40	Admitted with fever and left hypochondrial pain. Found to have enlarged tender spleen and left basal crepitations.	18.3 × 9
9	19	M	38	Admitted with left hypochondrial pain and fever. Found to have enlarged tender spleen and left basal crepitations.	15.2 × 7.8
10	20	M	38.8	Admitted with abdominal pain and fever. Developed salmonella septicemia. Found to have enlarged tender spleen.	15 × 7

sickling test and hemoglobin electrophoresis using Helena Laboratories Super Z Electrophoresis Kit (England). A drop of packed red blood cells was hemolyzed with hemolysate reagent applied to Titan III cellulose acetate plate and electrophoresed in Super Hemebuffer (pH 8.2 to 8.6) for 25 min. The plate was stained in Ponceau S, cleared, and the pattern quantified using Helena Quick Scan Densitometer. The number of Howell-Jolly bodies was measured and expressed per 1,000 RBC.

RESULTS

Among about 2,000 patients with SCD treated at our hospital, ten were treated for splenic abscess giving a 0.5% prevalence of splenic abscess in our SCD patients. There were 8 males and 2 females. Their ages ranged from 4 to 26 years (mean 16.5 years). Their relevant demographic and clinical data are shown in Table I. All of our patients presented with fever and abdominal pain and were found to have a tender enlarged spleen. Five of our patients were found to be septicemic on blood cultures. One of them was admitted 2 days after discharge from the hospital due to Enterobacter Sakazaki septicemia with a splenic abscess that grew Enterobacter Cloacae. The hematological parameters and cultures are shown in Table II. Their HbS ranged from 66.2–87 (mean 77.54) and their HbF ranged from 10.9–32.4 (mean 20.5). The WBC were elevated except in one patient (Table II). Five patients had thrombocytosis (normal platelet count = 140,000–440,000/ml) while four had thrombocytopenia and only one patient had normal platelets count. All four patients with thrombocytopenia had Salmonella septicemia proven by blood culture. Seven of our patients had increased Howell-Jolly bodies. One patient (Case 5) was found to have a large splenic abscess that had ruptured and extended subcutaneously. Three patients (Cases 6,8,9) also had left lower lobe pneumonia with pleural effusion in Case 9. Both CT and ultrasound demonstrated splenomegaly and irregular cystic areas within the splenic tissue with fluid suggestive of splenic abscess (Fig. 1). In one patient (Case 6) there was a large amount of fluid collection around the spleen, which was enlarged with a large hypoechoic area with fluid contents (Fig. 2). At operation, this was found to be a large splenic abscess that had ruptured. In two patients, it was difficult to differentiate between splenic abscess and a large splenic infarct. The diagnosis in these two patients was confirmed by needle aspiration under ultrasound guidance. In one patient (Case 7), a sulfar colloid technetium scan showed no uptake in the spleen.

All patients underwent splenectomy covered with antibiotics and after receiving 0.5 ml prophylactic pneumococcal vaccine (PMV-Immune 23 Lederle). One patient (Case 8) also had a cholecystectomy due to gall stones incidentally discovered intraoperatively, although preoperative ultrasound detected biliary sludge only. Case 6 also had an incidental appendecectomy. Case 7 had a splenectomy, appendecectomy, and distal pancreatectomy because of severe adhesions to the tail of the pancreas. A notable feature during surgery was the presence of dense adhesions between the spleen, anterior abdominal wall, diaphragm, left lobe of liver, splenic flexure of the colon, and greater omentum in most of the cases. The bacteriology of the abscesses is summarized in Table II.

Table II. Hematological Parameters and Cultures*

No.	HbS	HbF	HbA2	Hb	MCV	WBC	Platelets	HJB	Blood culture	Peritoneal and pus culture
1[a]	66.2	32.4	1.4	9.1	70	21.1×10^3	1,385,000	2/1,000	Enterobacter Sakazaki	Enterobacter Cloacae
2	69.5	28.9	1.6	9.6	61.9	17.3×10^3	48,000	1/1,000	Salmonella group D	N.G.
3[a]	77.9	19.7	2.5	7.9	71.9	21×10^3	132,000	3/1,000	Salmonella group D	N.G.
4	84.9	13.3	1.8	7.1	79.2	6.2×10^3	141,000	NO	Salmonella Spp.	N.G.
5	87	10.9	2.1	6.3	73.7	20.3×10^3	693,000	8/1,000	N.G.	Salmonella group D
6[a]	83	14.3	2.8	9.3	76.4	23.4×10^3	866,000	3/1,000	N.G.	Salmonella group D
7[a]	86.3	13.7	—	10.2	65.8	12.4×10^3	732,000	2/1,000	N.G.	N.G.
8	76.5	20	3.5	9.0	71.5	11.4×10^3	398,000	NO	N.G.	Enterobacter cloacae and Enterobacter Sakazaki
9[a]	77.1	20.6	2.3	8.1	82.3	24.5×10^3	667,000	5/1,000	N.G.	Coliforms
10[a]	67	30.8	1.9	10.6	75.9	12.2×10^3	96,000	NO	Salmonella group B	N.G.

*HJB = Howell-Jolly bodies; N.G. = no growth.
[a]Those with associated alpha thalassemia.

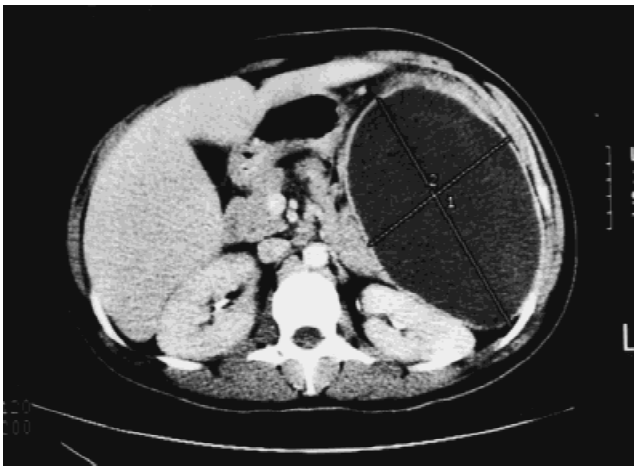

Fig. 1. CT-scan demonstrating a large splenic abscess.

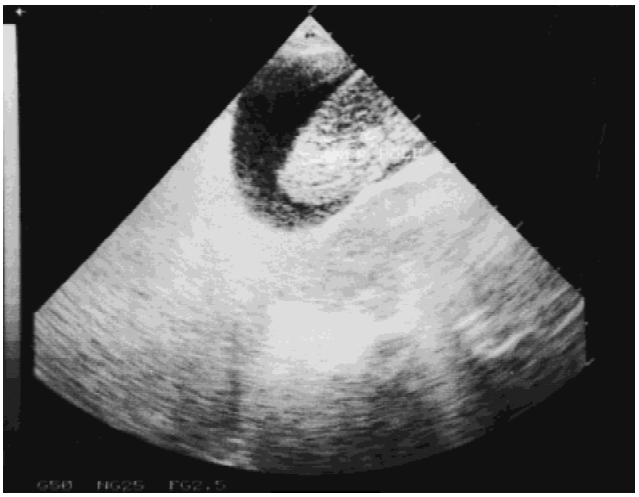

Fig. 2. Ultrasound demonstrating fluid collection around the spleen in a patient with ruptured splenic abscess.

Blood cultures were positive in 5 patients. Salmonella was isolated in 4 (80%) of them, while the pus cultures were positive in 5 patients, and two of them grew salmonella (40%). The histopathology of the resected spleens showed haemorrhage and engorgement of sinusoids by sickled RBC and large areas of infarction with abscess formation and hemosiderin deposition. In Case 5, there was cavitation and formation of a tract leading to splenic capsule and the abscess.

Post-operatively, all the patients recovered with no mortality. Two patients (Case 1 and 8) developed mild wound infection. Case 6 developed adhesive intestinal obstruction and Case 4 developed hematoma at the site of splenectomy, which required evacuation.

DISCUSSION

Splenic abscesses are known to be very rare in healthy individuals and are usually seen in patients with underlying pathology, which includes sepsis, splenic trauma with superimposed infection, spread from adjacent infectious process, or hemoglobinopathies [2]. In a review of 173 patients with splenic abscess, sepsis was the commonest predisposing factor in 73.4% of the cases, with infective endocarditis being the primary infectious disease most commonly associated with splenic abscess [2,15]. Hemoglobinopathies accounted for only 12% of the causes [2,15]. Splenic abscess, once considered to be uncommon, is being reported more recently. Immunodeficiency states due to disease or during immunosuppression have emerged as a new, common predisposing cause of splenic abscess [9–11].

Sickle cell disease is one of the commonly inherited hemoglobinopathies in the Eastern Province of Saudi Arabia and has been reported to be more benign than in other parts of the world [19–21]. This is attributed to high levels of HbF and the frequently associated alpha thalassemia [21–23]. In SCD, the spleen enlarges during the first decade of life, but then undergoes progressive atrophy and autosplenectomy due to repeated attacks of vasoocclusion and infarction, but sometimes splenomegaly persits into an older age group and, in some, into adult life. This is so in Saudi patients with SCD [22,24]. The reason for the failure of these individuals to undergo

autosplenectomy is unknown, but a correlation with persistent splenomegaly and a high level of HbF was established [25]. Our patients had a mean HbF of 20.5 (range 10.9–32.4) and 6 of them had associated alpha thalassemia. Persistence of splenomegaly in these patients predisposes them to the development of complications. One of these complications is splenic abscess. Splenic abscess in association with SCD was first described by Beet in 1949 [26]. Patients with SCD are known to be susceptible to systemic infections and one of the main reasons for this is the early development of functional asplenia in these patients [25]. At 3 years of age, 78% of SCD patients have elevated pocked RBC counts indicative of functional asplenia [27]. The increased susceptibility to infection and in the presence of splenic infarcts, which are not uncommon in patients with SCD, predisposes them to the development of splenic abscess [16–18].

Although the signs and symptoms of splenic abscess have been well described, they are not specific and so splenic abscess remains a diagnostic challenge. This is especially so in patients with SCD who are known to be febrile for other more common causes and frequently present with abdominal pain due to vasoocclusive crisis. Splenomegaly was observed in only 54% of 152 patients with splenic abscess [2,15], but we found a tender enlarged spleen of significance in patients with SCD. Splenic abscess should be considered in patients with SCD who present with fever and abdominal pain especially if found to have a tender enlarged spleen. These patients should be evaluated liberally by ultrasound and if in doubt by CT scan of the abdomen. Although ultrasound and CT scan are the best methods for the diagnosis of splenic abscess [28,29], we found CT scan more reliable and it allows more accurate anatomical localization of the abscess. Nelken et al. reported a 96% accuracy using CT-scan for the diagnosis of splenic abscess [29]. A high index of clinical suspicion is required for the early diagnosis of splenic abscess. This is to obviate the danger of splenic rupture. If not recognized and treated early splenic abscess may rupture locally, into the peritoneal cavity, into the adjacent bowel, or into the pleural space, leading to increased mortality [2,30,31]. Two of our patients presented with ruptured splenic abscess, one in the peritoneal cavity, and the other posteriorly to extend subcutaneously. A diagnostic difficulty in patients with SCD is the differentiation between splenic abscess and large splenic infarct, which can also present with a large tender spleen. This occurred in two of our patients and we found aspiration under ultrasound guidance of help in differentiating the two conditions.

Many organisms can cause splenic abscess but the most frequently encountered organisms are Staphylococci, Streptococci, and gram-ve bacilli [2,15]. In a review of 129 patients with splenic abscess, streptococci was the commonest organism followed by Staphylococci [2,15]. Salmonella accounted for 11% of the causative organisms [2,15]. Recently, however, other unusual organisms causing splenic abscess have been reported. These include pseudomonas [12,14], enterobacter [13], Serratia [8], non-typhoidal salmonella [32,33], and Klebsiella [30]. Fungi, which were once thought of as rare causes of splenic abscess, now together with mycobacterial are the commonest organisms causing splenic abscess in immunocompromized patients [9–11]. Two of our patients grew Enterobacter, but of interest was the isolation of salmonella in 2 of our patients from the abscess and in 4 from blood culture. Salmonella is known to cause various infections in patients with SCD including septicemia, osteomyelitis, and septic arthritis [34,35]. Although other organisms can cause splenic abscess in patients with SCD, we suggest preliminary antibiotic coverage including antibiotics against salmonella till culture and sensitivity results become available.

To obviate the dangers of total splenectomy especially the overwhelming postsplenectomy sepsis, splenic preservation is being increasingly advocated. This is so in the management of splenic abscess. Whereas the majority now recommend splenectomy for the management of splenic abscess, there are reports of partial splenectomy [8], CT-guided percutaneous catheter drainage [3–6], or even non-interventional treatment of splenic abscess [7]. Although percutaneous drainage under radiological guidance has been reported to be a safe and effective method for the treatment of splenic abscess both in children and adults [3–6], most authors recommend it for patients who are critically ill with unilocular abscess or in young patients to preserve their spleen [1,15]. We believe that the treatment of choice of splenic abscess in SCD patients is splenectomy and antibiotics. Percutaneous abscess drainage in the presence of personnel experienced in this technique can be used temporarily in patients who are sick and at risk from splenectomy, as there is no point in preserving a nonfunctioning spleen, which is present in the majority of these patients. In a review of 173 patients with splenic abscess, the overall mortality was 39.3% and for those treated with splenectomy it was 7% [2,15]. Our patients, a high-risk group, were treated successfully with splenectomy and antibiotics with no mortality and minimal morbidity.

REFERENCES

1. Herman P, Oliveira E, Silva A, Chaib E, D'albuquerque LC, Pugliese V, Machado MCC, Pinotti HW: Splenic abscess. Br J Surg 82:355, 1995.
2. Chun CH, Raff MJ, Contreras L, Varghese R, Waterman N, Daffner R, Melo JC: Splenic abscess. Medicine 59:50–65, 1980.
3. Berkman WA, Harris SA Jr, Bernardino ME: Non-surgical drainage of splenic abscess. AJR 141:395–396, 1983.
4. Lerner RM, Spataro RF: Splenic abscess: Percutaneous drainage. Radiology 153:643–645, 1984.
5. Gerzot SG, Robbins AH, Johnson WC, Birkett DH, Nabseth DC:

Percutaneous catheter drainage of abdominal abscesses. N Engl J Med 305:653–657, 1981.
6. Ramakrishnan MR, Sarathy TKP, Balu M: Percutaneous drainage of splenic abscess: Case report and review of the literature. Pediatrics 79:1029–1030, 1987.
7. Fernandes ET, Tavares PB, Garcette CBM: Conservative management of splenic abscess in children. J Pediatr Surg 27:1578–1579, 1992.
8. Bhattacharyya N, Ablin DS, Kosloske AM: Stapled partial splenectomy for splenic abscess in a child. J Pediatr Surg 24:316–317, 1989.
9. Hatley RM, Donaldson JS, Raffensperger JG: Splenic microabscesses in the immuno-compromized patient. J Pediatr Surg 24:697–699, 1989.
10. Marmon LM, Vinocur CD, Wimmer RS, Konefal SH, Weintraub WH: Fungal splenic abscesses: Management in childhood leukemia. Pediatr Surg Int 5:118–120, 1990.
11. Smith MD Jr, Nio M, Camel JE, Sato JK, Atkinson JB: Management of splenic abscess in immunocompromized children. J Pediatr Surg 28:823–826, 1993.
12. Ooi LLPJ, Nambiar R, Rauff A, Mack POP, Yap TL: Splenic abscess. Aust NZJ Surg 62:780–784, 1992.
13. Cheema MA, Al-Saigh A, Abdul Latif A: Splenic abscess: Presentation, diagnosis and management. Ann Saudi Med 12:552–554, 1992.
14. Studemeister AE, Beilke HA, Kirmani N: Splenic abscess due to clostridium difficile and pseudomonas paucimobilis. Am J Gastroenterol 82:389–390, 1987.
15. Gadacz TR: Splenic abscess. World J Surg 9:410–415, 1985.
16. Cavenagh JD, Joseph AEA, Dilly S, Bevan DH: Splenic sepsis in sickle cell disease. Br J Haematol 86:182–189, 1994.
17. Al-Salem AH, Mallapa KK, Qaisaruddin S, Al-Jam'a A, Elbashier A: Splenic abscess in children with sickle cell disease. Pediatr Surg Int 9:489–491, 1994.
18. Grant CS, Al-Salem A, Khwaja MS, Sumer T, Al-Awamy B: Splenic abscess in children: Aspects of management. JR Coll Surg Edinb 32: 342–345, 1987.
19. Al-Awami BH: Sickle cell anemia: Its clinical manifestations and their management. Saudi Med J 8:553–562, 1987.
20. Al-Awami BH, Niazi GA, El-Mouzan MI, Altorki MT, Naeem M: Newborn screening for sickle hemoglobinopathies and other inherited erythrocytic disorders in the eastern province of Saudi Arabia. Saudi Med J 7:502–509, 1986.
21. Perrine RP, Pembrey ME, John P, Perrine S, Shoup F: Natural history of sickle cell anemia in Saudi Arabia. Ann Intern Med 88:1–6, 1978.
22. Perrine RP, Brown MJ, Clegg JB, Weatherall DJ, May A: Benign sickle cell anemia. Lancet 2:1163–1167, 1972.
23. Al-Awami B, Niazi GA, El-Mouzan MI, Altorki MT, Naeem M: Relationship of hemoglobin F and alpha-thalassemia to severity of sickle cell anemia in the eastern province of Saudi Arabia. Ann Trop Pediatr 6:261–265, 1986.
24. Al-Salem AH, Qaisaruddin S, Nasserullah Z, Al Dabbous I, Abu Srair H, Al Jam'a A: Splenectomy and acute splenic sequestration crisis in sickle cell disease. Pediatr Surg Int 11:26–28, 1995.
25. Pearson HA, Gallagher D, Chilcote R, Sullivan E, Wilimas J, Espeland M, Kim Ritchey A: Developmental pattern of splenic dysfunction in sickle cell disorders. Pediatrics 76:392–397, 1985.
26. Beet EA: Primary splenic abscess and sickle cell disease. East Afr Med 22:180–186, 1949.
27. Brown AK, Sleeper LA, Miller ST, Pejelow CH, Gill FM, Waclawiw MA: Reference values and hematologic changes from birth to 5 years in patients with sickle cell disease. Arch Pediatr Adolesc Med 148: 796–804, 1994.
28. Ralls PW, Quinn MF, Colletti P, Lapin SA, Halls J: Sonographic of pyogenic splenic abscess. Am J Roentgenol 138:523–525, 1982.
29. Nelken N, Ignatius J, Skinner M, Christensen N: Changing clinical spectrum of splenic abscess: A multicenter study and review of literature. Am J Surg 154:27–34, 1987.
30. Alloush NM, Alloush N: Unusual presentation of splenic abscess. Saudi Med J 12:148–149, 1991.
31. Lawhorne TW, Zuidema GD: Splenic abscess. Surgery 79:686–689, 1976.
32. Rodan BA, Max Rj, Breiman RS, Rice RP: Splenic abscess due to salmonella typhimurium bacteremia. South Med J 74:382–383, 1981.
33. Debeuckelaere S, Schoors DF, Buydens P, DuVille L, Spapen HDM, Reynaert HM, DeVis G: Splenic abscess: A diagnostic challenge. Am J Gastroenterol 86:1675–1678, 1991.
34. Al-Salem AH, Ahmed HA, Qaisaruddin S, Al-Jam'a A, Elbashier AM, Al-Dabbous I: Osteomyelitis and septic arthritis in sickle cell disease in the eastern province of Saudi Arabia. Int Orthopaed 1:398–402, 1992.
35. Elbashier AM, Abusrair H, Al-Jam'a AH, Al-Dabbous IA, Malik AG: Septicemia in sickle cell disease patients at Qatif Central Hospital, Saudi Arabia. Saudi Med J 13:220–223, 1992.